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Case Report

Malignant peripheral nerve sheath tumor of the nose with coincidental HIV infection: Management challenges in limited resource setting

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ABSTRACT

Malignant peripheral nerve sheath tumor (MPNST) is an uncommon soft-tissue sarcoma, with origin from the peripheral nerves or somatic tissues associated with nerve sheath. This tumor rarely affects the nose. The clinicopathological presentations vary from one case to another and depend on cells of origin. Hence, the diagnosis is often difficult. We report a 36-year-old woman who presented in our facility with progressive nasal swelling, facial deformity, and mucopurulent rhinorrhea of 3 years duration. Examination revealed a significant facial disfigurement. She was also found to be seropositive. Computerized tomography scan of paranasal sinuses showed an expansile, subtle enhancing, predominantly low-density mass, with area of air density within it, completely occupying the nasal cavity. No calcific focus, lytic or sclerotic surrounding bones changes seen. Her tumor was resected under general anesthesia with a wide margin, and regional vital structures were preserved. The histological/immunohistochemistry revealed MPNST. She had adjuvant radiotherapy and chemotherapy 2 weeks after surgery. However, a nodule was seen on the floor of the nose few days after completion of radiotherapy, which was biopsied, and histological report was positive for malignancy. The persistence of the malignancy and HIV hampered her facial reconstruction. Also, limited finance hampered procurement of the nasal prosthesis. She was followed up for 1 year before she was lost.

Keywords: Peripheral nerve, Nose, HIV infection, Soft tissue, Sarcoma

INTRODUCTION

Malignant peripheral nerve sheath tumor (MPNST) is also known as neurogenic sarcoma, malignant neurilemoma, malignant schwannoma, and neurofibrosarcoma.[1] It arises from a peripheral nerves or displays differentiation along various elements of nerve sheath. It may originate from multiple cell types, namely, Schwann cells, perineural fibroblasts, or fibroblasts.[2]

MPNST is a rare sarcoma. Correspondingly, a MPNST of the head and neck is very rare and even rarer when is located in the nasal cavity and paranasal sinuses.[1,3,4] The primary MPNST of the nose and paranasal sinuses, and head and neck accounts for only 4% of all neural tumors of the head and neck.[1]

The two main risk factors associated with MPNST are neurofibromatosis type 1 (NF1); account for about 50–60% of cases,[5] and radiotherapy which accounts for 10%.[3] However, some cases occur sporadically.[5]

Diagnosis could be quite challenging especially in sporadic cases. There are no specific clinical features for MPNST; also radiological imaging studies cannot effectively differentiate benign from malignant lesions.[6]

Diagnosis is, therefore, based on tissue biopsy for histological and immunohistochemical features of Schwannian differentiation.[7]

Prognosis of this lesion depends on the tumor size, histological grade, the resection margin, metastasis, and anatomical location of the tumor.[5,8] MPNST has propensity for local invasion, recurrence, and distant metastasis to the lungs and peritoneum,[3] hence carries a poor prognosis. Furthermore, location of MPNST in nasal cavity portends worse prognosis.[4]

The main stay of therapy is radical resection with adequate surgical safe margins of this tumor.[5] However, some patients may require adjuvant radiotherapy with or without chemotherapy.

We present a case report of sporadic MPNST of external nose with coincidental HIV in a 36-year-old woman. She had wide resection of the tumor with adjuvant radiotherapy and chemotherapy. Reconstructive surgery of her nose could not be done because of discovery of malignant nodule on the floor of the nose 15-day post-radiotherapy. Persistent malignancy, HIV infection, and extensive facial defect were the poor prognostic factors; and they constituted to management challenges in this case. The limited resource was another challenge that made it impossible for her to procure recommended prosthesis.

CASE REPORT

A 36-year-old woman, a cassava farmer, was brought to our clinic in 2016 by a missionary with 3-year history of insidious onset of a growth on the right side of nasal bridge that was initially about size of a peanut, but progressively increased in size and spread to the entire external nose. There was headache, biting sensation on the nose, and thick yellowish foul-smelling blood stained mucopurulent rhinorrhea, anosmia, and weight loss. There were no associated fever, cough, night sweat, hoarseness, and no visual defect. She had no history of atopy or nasal trauma. She was not a known diabetic (fasting blood sugar was 3.5 mmoI/L). Incidentally, she was found to be HIV seropositive on admission.

General physical examination revealed a chronically ill woman with huge disfigured multinodular nasal mass about 9 × 7 × 6 cm with ulcerated tip of the nose [Figures 1a and 1b]. The mass is fixed to both skin and underlying structures. All other systems were within normal limit.

Biochemical parameters were with normal limit except bilirubin total (21.5 umol/L and conjugated (12.5 umol/L). Total white blood cell count was 3.7×10^9/L, neutrophil was 55%, eosinophil; 5%, and lymphocytes; 40%. X-ray chest was normal. Computed tomography (CT) scan of paranasal sinuses showed an expansile, subtle enhancing, predominantly low-density mass, with area of air density within it, completely occupying the nasal cavity. It measured 6.55 cm × 7.13 cm × 7.48 cm in its widest perpendicular dimensions. No calcific focus, lytic or sclerotic surrounding bones changes seen. The surrounding paranasal sinuses were clear. The orbit and its contents were within normal limits. Postnasal space and the remaining air passages were preserved. Other surrounding structures were normal.

Histological report of punch biopsy of the lesion showed MPNST [Figure 1c]. She gave her consent for both surgical resection of the tumor and post-operative radiotherapy. The tumor resection was done under general anesthesia through endotracheal intubation. Tissue removed was subjected to histological examination, which revealed MPNST [Figures 1d] and confirmed by immunohistochemistry, which showed positive reaction for S-100 protein [Figures 1f and 1g]. She commenced radiotherapy 2 weeks after operation and received a total dose of 6000 cGy in 30 daily fractions over 6 weeks on Cobalt 60 machine. The procedure was well tolerated [Figure 1h]. She was also placed on HAART for HIV infection in addition for treatment received for MPNST. Subsequently, she was referred to the plastic surgeons for nasal reconstruction. Unfortunately, the plastic surgeons discovered a nodule on the floor of the nose 15 days after radiotherapy. This nodule was biopsied and reported positive for malignancy [Figure 1e]. Because of the persistent MPNST and HIV status, the plastic surgeons could not reconstruct her nose but rather recommend prosthesis. The extensive defect on her face couple with difficulty in accessing prosthesis because of limited resource was our patient challenge. She was followed up for 1 year, thereafter defaulted and reported dead at home 2 years later.

DISCUSSION

MPNST is a rare neoplasm. It occurs in all ages but peaks at the second and fourth decades,[1] however, the patients with NF1 often present at younger age than the sporadic and post-radiation counterparts.[7] It has no predilection for sex and race.[1]

MPNST may be a malignant transformation of pre-existing neurofibroma or neurofibromatosis, may occur due to previous radiotherapy, or may be sporadic in origin.[1,7,9] MPNST is most commonly located on the lower extremities[10] and retroperitoneum,[3] and its extremely rare in the nose and paranasal sinuses.[1] The most common site in nose and paranasal sinuses is the ethmoid and maxillary sinuses. It can also occur in the nasal cavity and sphenoid sinus. The least site of occurrence is the frontal sinus.[1]

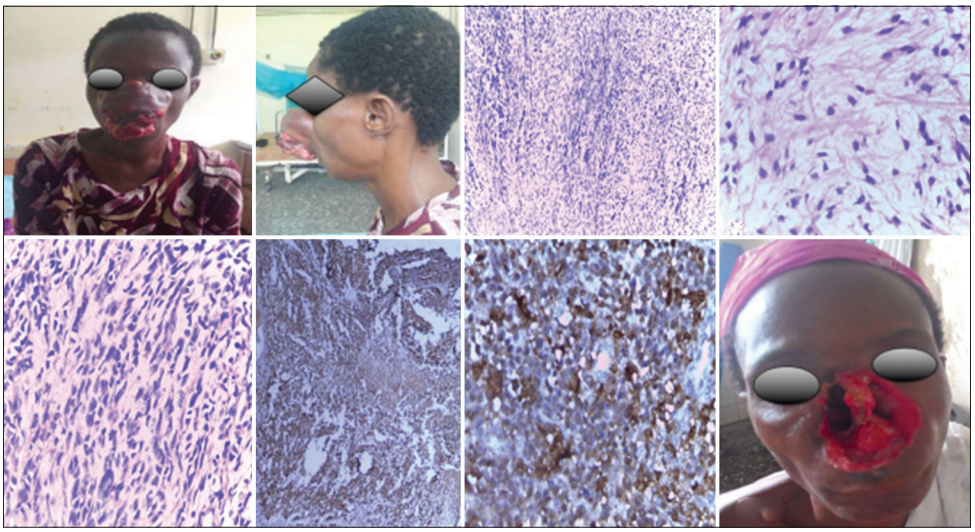

Figure 1a-h: A 36-year-old woman with MPNST of the external nose presented with significant facial disfigurement. The photographs: anteroposterior view (Figure 1a) and left lateral view (Figure 1b) at presentation. Photomicrograph of histologic section shows both alternating cellular and hypocellular areas (Figure 1c; H and E ×100). Photomicrograph of histologic section of hypocellular area shows a myxoid stroma with scanty malignant spindle shape cells with bizarre looking nuclei and coarse chromatin pattern (Figure 1d; H and E ×400). Photomicrograph of histologic section shows cellular area consisting malignant spindle shape cells with bizarre looking nuclei and coarse chromatin patterns and ovoid-shaped tumor cells with tumor giant cells (Figure 1e; H and E ×400). Photomicrograph shows strong immunoreactivity for S-100 protein (Figure 1f and 1g). The photograph post-radiotherapy (Figure 1h).

The origin of MPNST has been linked with multiple cell types; such as Schwann cells, perineural cells, or fibroblasts; hence, the clinicopathologic features differ from one case to another with resultant diagnostic difficulty.[3] Often times, MPNST presents insidiously and diagnosis may be delayed or missed.[10] Clinical features of MPNST of the nose and paranasal sinuses include unilateral nasal obstruction, epistaxis, facial pain, swelling of the facial and orbital region, mucopurulent rhinorrhea, hyposmia, and headache.[1] Unfortunately, these clinical features are not specific for MPNST alone. Other pathological lesions that may present with same symptoms and signs are nasal polyps, mucocele, gliomas, papillomas, esthesioneuroblastomas, sarcomas, carcinomas, and lymphomas. However, MPNST is characterized by local aggression and erosion of adjacent bone and soft tissue, multiple recurrence, and metastases by hematological route to lungs and visceral.[1,3] MPNST does not have specific characterizations on anatomical radiological images.[9] Nevertheless, the role of radiological images such as CT scan and MRI is to help in staging the malignancy and monitor the disease progression.[1]To reach the definitive diagnosis, tissue biopsy is mandatory for histology and immunohistochemistry.

The tissue biopsy histological report of our case report was MPNST, which was confirmed by immunochemistry. The index patient was a middle-aged woman with sporadic MPNST of the external nose. This case is unique because of coincidental HIV infection with MPNST. She is the first reported case of MPNST in this geopolitical region of our country and supposedly second reported case in a Black African. She had no pre-existing neurofibroma, family history of von Recklinghausen disease, or previous history of radiotherapy. The tumor invaded the soft tissue of the entire nose without bony invasion of the surrounding structures. There was no metastasis to the lungs.

The main stay of treatment for MPNST is complete surgical excision of the tumor with safe surgical margin. Our patient's tumor affected the entire external nose with involvement of both the skin and underlying structures and ulceration of some parts of the tumor. The tumor was resected with wide margin, preserving the surrounding vital structures. Subsequently, she had post-operative radiotherapy and chemotherapy. Furthermore, she was treated for HIV infection with HAART. The reconstructive surgery schedule could not be carried out because of persistent malignancy and HIV infection. She was, therefore, offered prosthesis. Unfortunately, prostheses could not be obtained because of paucity of funds and non-availability of this product in our environment.

CONCLUSION

This case is unique because of coincidental HIV with MPNST and management was quite challenging. We commended the missionary that took care of her despite their meager resource.

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Authors' contribution

AAG, substantial contribution to conception and design of the work, descriptive and reporting, drafting the work and revising it critically for important intellectual content, final approval of version to be published, and agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved; UTI conception and design, descriptive and reporting, manuscript drafting, critical revision of manuscript, final approval of revision to be published, and agreement to be accountable; MRB conception and design, descriptive and reporting, manuscript drafting, critical revision of manuscript, final approval of revision to be published, and agreement to be accountable; and KGA conception and design, descriptive and reporting, manuscript drafting, critical revision of manuscript, final approval of revision to be published, and agreement to be accountable.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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